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Spontaneous Page kidney from ruptured renal angiomyolipoma

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ABSTRACT

Renal angiomyolipomas are the most common benign tumors of the kidneys. They are prone to rupture, which may result in massive hemorrhage and often requires lifesaving nephrectomy. Delay in treatment is likely to result in death. We report two cases of ruptured angiomyolipoma compressing the renal parenchyma, causing secondary hypertension (Page kidney). Both patients presented with abdominal pain, hypertension, and reduced or dropping hemoglobin counts. The delay in diagnosis and treatment resulted in their adverse outcomes. We highlight the need to promptly diagnose and treat symptomatic renal hematomas to avoid subsequent morbidity and mortality.

KEYWORDS Acute kidney injury; angiomyolipoma; Page kidney; spontaneous perinephric hematoma

Page kidney occurs due to extrinsic compression of the renal parenchyma by a mass or hematoma, leading to activation of the renin-angiotensin-aldosterone system, causing secondary hypertension.[1] Common causes are renal tumors including angiomyolipoma, trauma especially from sports, motor vehicle accidents, iatrogenic causes, anticoagulation, polyarteritis nodosa, operative, and idiopathic.[2] Renal angiomyolipomas (RAMLs) are the most common benign tumors of the kidneys. The most common and life-threatening complication of angiomyolipoma is rupture and subsequent hemorrhage.[3]

CASE DESCRIPTIONS

The first case was a 55-year-old hypertensive, diabetic woman who was admitted with 1 week of abdominal pain, cough, fever, dyspnea, and elevated blood pressure. Ultrasound of the abdomen revealed previously seen benign right kidney angiomyolipomas (largest mass of 0.9 cm). She was diagnosed with acute respiratory failure from COVID-19 pneumonia and treated with remdesivir, dexamethasone, and enoxaparin. She continued to experience abdominal pain. On hospital day 14, her hemoglobin dropped to 2 g/dL, prompting computed tomography (CT) of the abdomen, which revealed a large right perinephric expanding hematoma that compressed the right kidney *(Figure 1a, 1b)*, for which the main and accessory renal arteries were embolized. Hemodialysis was initiated, and she died from complications. Autopsy revealed necrosis due to embolization at the site of renal hematoma corresponding to prior larger angiomyolipomas *(Figure 1c)*.

The second case was a 42-year-old man with hypertension who presented with 3 weeks of diffuse right lower abdominal pain and elevated blood pressure. He was seen at another hospital a week earlier for scrotal pain and swelling, which was diagnosed as right-sided hydrocele on scrotal ultrasound. During the current presentation, his right lower abdominal pain was indolent, sharp, colicky, and associated with fatigue and intermittent dizziness without a history of trauma, fever, diarrhea, hematuria, or melena. His blood pressure was 189/127 mm Hg; he had tenderness and guarding throughout the right lower abdomen and had a mildly edematous right-sided scrotum. Abdominal CT showed a large right perinephric and subcapsular hematoma without active extravasation, confirmed by CT angiography of the abdomen *(Figure 2)* with a 2.4 cm lesion, suspicious for

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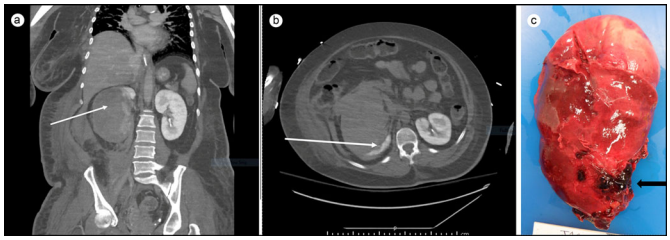

Figure 1. Case 1. Computed tomography angiogram of the abdomen—**(a)** coronal view and **(b)** axial view—revealing large right perinephric expanding hematoma with active extravasation, compressing the right kidney (arrow). **(c)** Autopsy sample, with lower pole showing the site of necrosis after angioplasty corresponding to the site of angiomyolipoma (arrow).

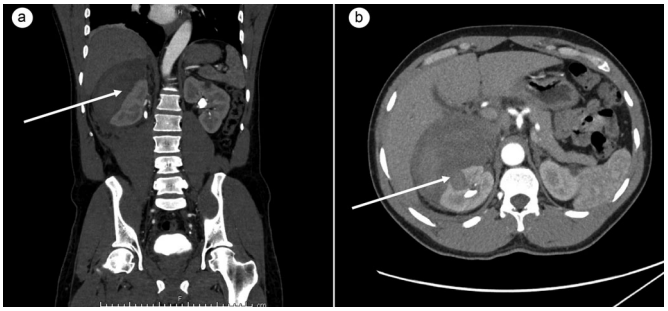

Figure 2. Case 2. Computed tomography angiogram of the abdomen—**(a)** coronal view and **(b)** axial view—showing a right perinephric and subcapsular hematoma with a small, contiguous lesion within the anterior right renal parenchyma (arrow).

angiomyolipoma. A renal angiogram showed no active extravasation or pseudoaneurysm. Interventional radiologists conducted particle embolization of the suspected angiomyolipomatous lesion. On follow-up in the urology clinic, his hemoglobin had improved and scrotal swelling had resolved. Features of the two cases are summarized in *Table 1*.

DISCUSSION

RAMLs account for 3% of all solid renal masses,[4] with a prevalence of 0.13% in the general population. Sporadic RAML is the most common. Spontaneous ruptures can result in nephrectomy, hemorrhagic shock, or even death.[4] Scoring systems have been established based on clinical features for prediction of RAML rupture and hemorrhage. RAML with flank pain on the affected side was highly associated with tumor rupture and hemorrhage. Tumors >4 cm were most likely to rupture and require continued surveillance.[4] In our patients, although the RAML was <4 cm, the clinical features of same side flank and abdominal pain, nonresolving scrotal pain, sudden onset anemia, and hypertensive emergency were indicative of rupture of RAML causing Page kidney. Prompt radiological evaluation would have helped diagnose these conditions sooner and allowed for lifesaving interventions. In addition, the use of therapeutic anticoagulation to treat hypercoagulability with COVID-19 in case 1 posed a risk of bleeding in the angiomyolipomas.[1] Prompt withholding of anticoagulation may have reduced the morbidity with COVID pneumonia and avoided our patient's eventual demise.

Table 1. Clinical observations in the two cases of Page kidney

Variable	Case 1	Case 2
Age (years)	55	42
Sex	Woman	Man
Body mass index (kg/m^2)	44.3	27.7
Previous stroke	0	+
Hypertension	+	+
Diabetes mellitus	+	0
Chronic kidney disease	0	+
Admission blood pressure (mm Hg)	178/81	189/127
Abdominal pain	+ (moderate intensity)	+ (indolent)
Duration of symptoms (weeks)	1	4
Other symptoms	Fever, cough, dyspnea, COVID-19 pneumonia	Fatigue, intermittent dizziness, right scrotal swelling
Possible precipitating feature	Enoxaparin*	Spontaneous
Urinalysis: Proteinuria	+	+
Urinalysis: blood present	+	+
Lesion size (cm)	0.9	2.4
Hemoglobin (g/dL) before imaging	15.5 → 7.8 → 2.0	8.4 → 8.9 → 10.7
Serum creatinine (g/dL) before imaging	0.5 → 2.3	1.4 → 1.3
Perinephric hematoma	+	+
Acute kidney injury	+	0
Need for dialysis	+	0
Renal embolization	+	+
Died	+	0

*1 mg/kg for COVID-19–induced hypercoagulopathy.

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